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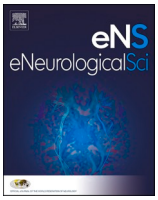

Adding a chapter to the literature: A rare encounter of unilateral Moyamoya disease with ipsilateral persistent trigeminal artery

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ABSTRACT

Background: The coexistence of persistent trigeminal artery (PTA) and Moyamoya disease (MMD) has been reported. If their pathogenesis is related and if PTA is protective or harmful in MMD remains unknown as these are rare cerebrovascular anomalies.

Case presentation: A 35-year-old woman with sudden global aphasia whose CT head and CT angiography of head and neck showed a hypodensity in the left posterior middle cerebral artery (MCA), a possible left proximal internal carotid artery occlusion, and a left PTA with hypoplasia of vertebral and basilar arteries. Digital subtraction angiography showed chronic MMD in the left MCA with extensive pial collateralization from anterior cerebral artery (ACA). The patient was initiated on single antiplatelet therapy and later she underwent direct bypass surgical intervention and rehabilitation.

Discussion: Our case report brings attention to the infrequent coexistence of ipsilateral MMD and PTA suggesting a potential congenital pathogenesis based on embryologic development and hemodynamics. Also, we propose a protective role of PTA in MMD in case of large anterior vessel occlusion. This case contributes to the scarce literature on the intriguing relationship between MMD and PTA.

1. Introduction

The coexistence of persistent trigeminal artery (PTA) and Moyamoya disease (MMD) has been reported in the past [1–6]. To understand the significance of this finding it is important to review its origins.

During the early embryo formation (4-5 mm) the forebrain is supplied by the carotid system, and the hindbrain by two parallel longitudinal neural arteries that obtain their blood supply from four carotid-vertebrobasilar anastomoses named trigeminal, otic, hypoglossal, and proatlantal arteries. [7–9] Later, at the 11–12 mm embryological stage, the medial cerebral artery (MCA) starts to develop as small buds on the anterior division of the primitive internal carotid artery (ICA). [8] Is also during this period, 5- to 14-mm embryo stage, that the basilar artery forms making these anastomoses to be obliterated. [9] The trigeminal artery is the last of the carotid vertebrobasilar anastomoses to be obliterated and is the most common to persist with a prevalence of 0.06–2.2%. [10]

The etiology of Moyamoya disease is unknown but studies proposed it originates from a problem in the angiogenesis that prevents the fusion of primitive small vessels. [5] If the formation of MMD and PTA are related and if PTA is protective or harmful in MMD remains unclear as these are rare cerebrovascular anomalies. We aim to improve the understanding and management of rare cerebrovascular anomalies. This case shows the thorough evaluation and comprehensive management of a patient with unilateral MMD, PTA and hypoplastic vertebral and Basilar arteries.

2. Case

A 35-year-old woman with history of essential hypertension and polysubstance abuse (cocaine), presented to the Emergency after a sudden onset of global aphasia, last known well was six hours before arrival. Her blood pressure was 197/120, her NIHSS score was 6 (questions 2, perform tasks 2, language 2). The Computed Tomography of head (CTH) showed an area of hypodensity in the left parieto-occipital lobe (Fig. 1) and the Computed Tomography angiogram of head and neck showed a possible left terminal ICA occlusion, with diminutive filling of the left MCA, also a left PTA with hypoplasia of vertebral and basilar arteries (Fig. 2.A). She was out of the time window for thrombolysis, but a potential candidate for thrombectomy. Digital subtracting angiography showed signs of chronic MMD in the left MCA with extensive pial collateralization from the anterior cerebral artery (ACA) (Fig. 2.B). We started therapy with a single antiplatelet and high dose statin, and hypertonic saline. Magnetic resonance imaging (MRI) showed an acute stroke involving the left posterior MCA and left posterior cerebral artery (PCA) territory (Fig. 3). On the second day's evaluation she was somnolent, with expressive aphasia and right homonymous hemianopia. She was transferred to Neurology Intensive care unit for medical management of cerebral edema, with great clinical improvement. Patient underwent surgical bypass revascularization a week after. On her immediate post op she developed a sudden left sided gaze, right sided weakness in face and body, and global aphasia. New MRI showed acute left posterior MCA ischemic strokes. She was started on a single antiepileptic drug and continuous electroencephalogram for 3 days which did not show seizures. She was later discharged with moderate expressive aphasia and mild right face and upper extremity

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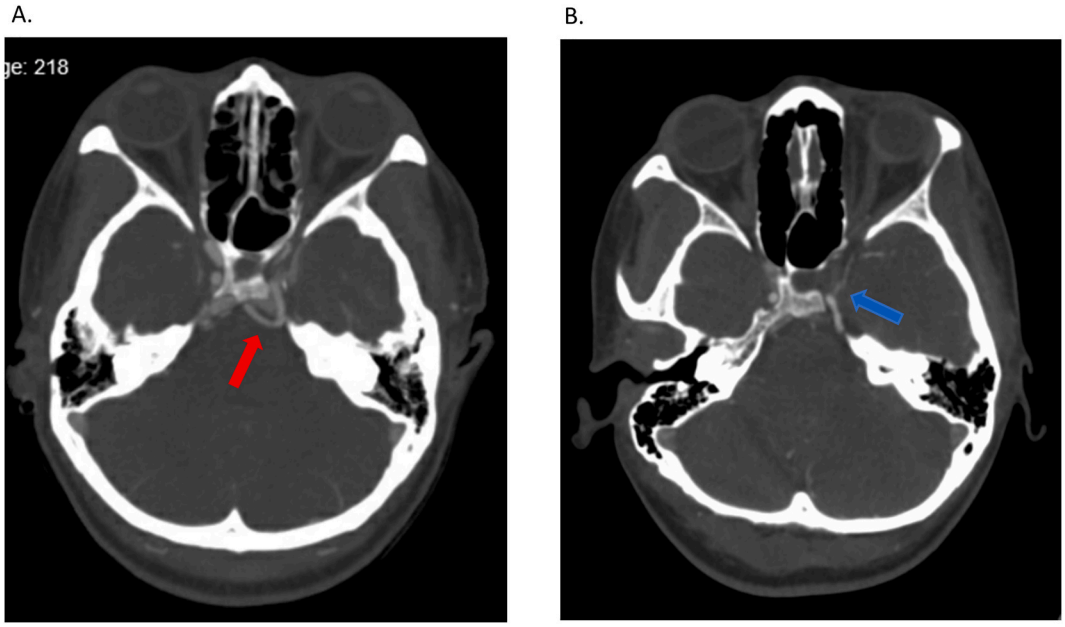


Fig. 1. Computed tomography angiogram of head and neck. **A.** Axial view shows a left persistent trigeminal artery (red arrow). **B.** Axial view of the left persistent trigeminal artery supplying blood to its ipsilateral medial cerebral artery (blue arrow).

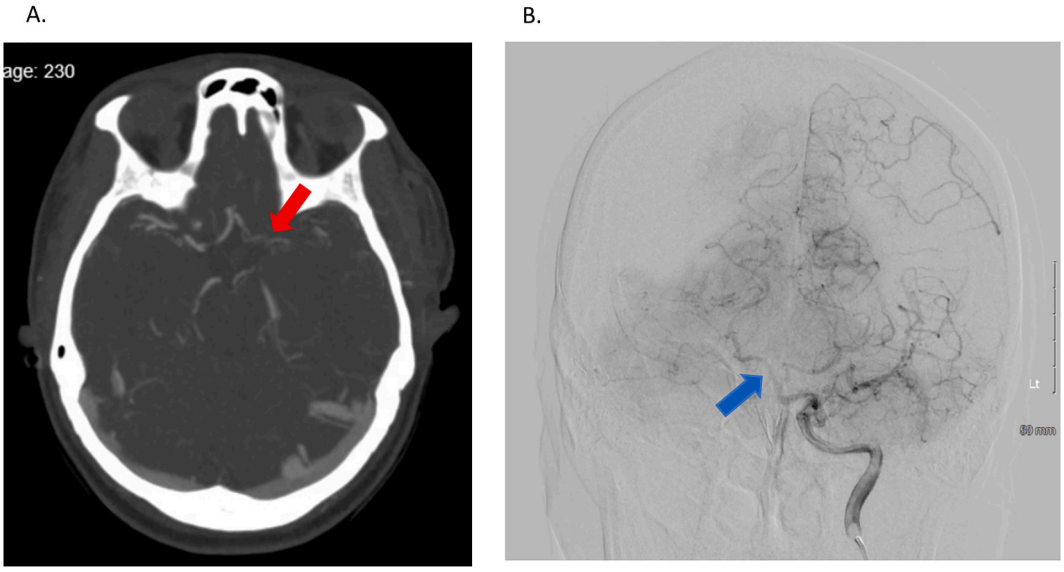


Fig. 2. **A.** Computed tomography shows limited filling of left medial cerebral artery (red arrow) and **B.** Digital subtracting angiography shows cross-filling through the right anterior cerebral artery (blue arrow) to left anterior cerebral artery and left medial cerebral artery.

weakness. On phone follow-up two weeks post-surgery her expressive aphasia is improving, she can feed herself with a spoon, and ambulate without a device, and follows up with speech and physical therapy.

3. Discussion

The embryologic angiogenesis is the growth of new vessels from pre-existing ones, and its main mechanism is sprouting. [11] As more capillaries are formed, the resistance to flow is reduced in larger arteries facilitating remodeling of the arteries. [11] During the 5- to 6-mm embryo stage the posterior communicating artery (P-comm) develops and connects with the distal basilar artery (BA), making the carotid and vertebrobasilar arteries anastomoses to regress. (Fig. 4) [12]. The development of the MCA is first identified as small buds in the primitive ACA by the 11–12 mm embryological stage. [7]

Many case reports documented coincidental findings of MMD and PTA [1–6] and explore the possibility of a common pathogenesis. [2,5] Nielsen and Johnson found that stenosis of the carotid caused filling of the PTA by vertebral angiography and the blood flow of the vertebrobasilar system was directed into the carotid system through the PTA. (Nielsen and Johnson 1967) Based on the chronological formation of the brain arteries and the hemodynamics role in artery remodeling, we hypothesize that in case of hypoplasia of P-comm and basilar artery, the trigeminal artery persists, diverging the blood flow from the carotid artery during the MCA formation, promoting MMD.

Besides MMD and PTA pathogenesis, studies have also discussed if PTA results protective or harmful in MMD [2,5,13], in our case of unilateral ipsilateral MMD the PTA plays a protective role because in case of

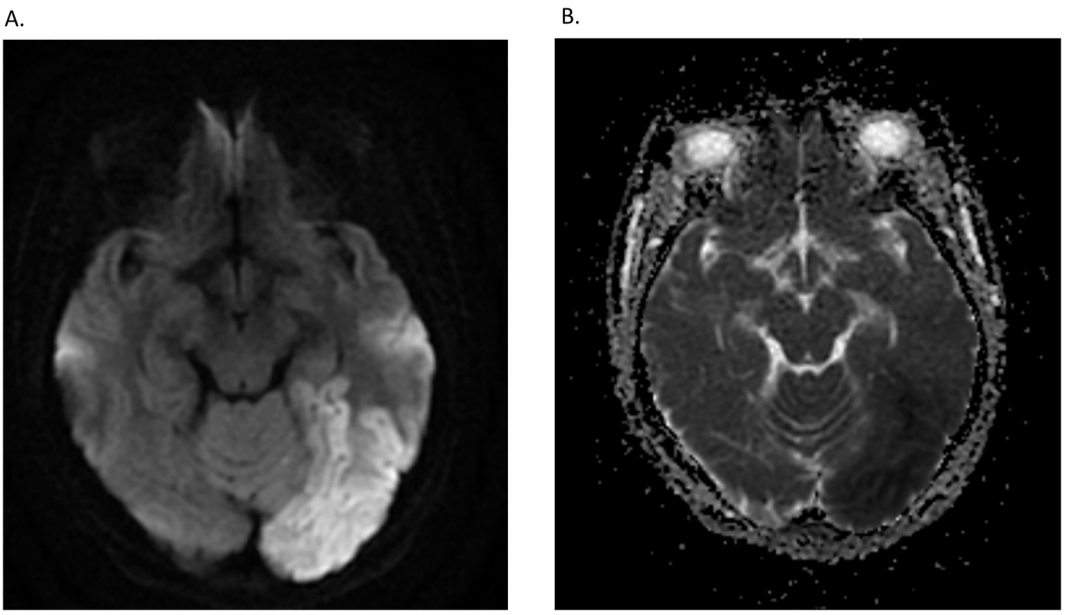


Fig. 3. A. Magnetic Resonance Imaging showed Diffusion Restriction in left parieto-occipital area with B. apparent diffusion coefficient correlation representing acute stroke.

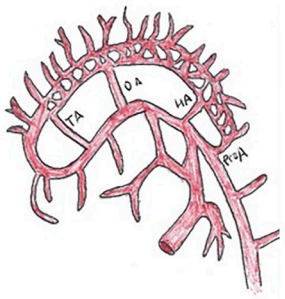
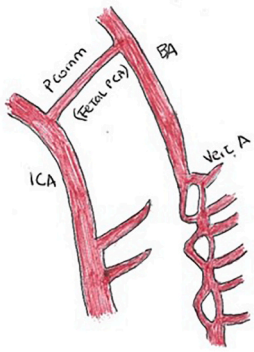
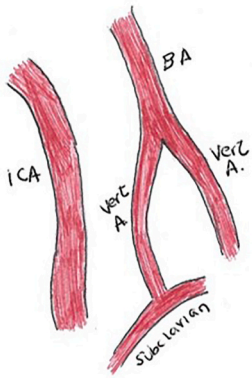
Padget Stage 2	Padget Stage 3	Padget Stage 4
		
4-5 mm stage	7-12mm stage	12-14mm stage
TA Trigeminal artery. **OA** Otic artery. **HA** Hypoglossal artery. **ProA** Proatlantal artery.	**ICA** Internal carotid artery. **Pcomm** Posterior communicating artery. **BA** Basila artery. **Vert A** Vertebral artery	

Fig. 4. Phases of development of posterior circulation. Initially the posterior circulation blood supply comes from the anterior circulation through carotid-vertebrobasilar anastomoses and later the posterior communicating artery and Basilar artery forms, causing obliteration of the anastomoses from caudal to rostral.

anterior large vessel occlusion the PTA would provide blood supply to the posterior circulation.

The unilateral MMD and history of polysubstance abuse raised questions regarding if this was a case of Moyamoya syndrome. The common causes of Moyamoya Syndrome (MMS) are arteritis, radiation injury, trauma, cocaine abuse, arteriosclerosis, leptospirosis infection. [14] The database search of MMS and cocaine use resulted in a few cases that reported bilateral anterior circulation involvement, but none with unilateral. [14–17] The findings of unilateral Moyamoya, hypoplastic posterior circulation and PTA incline this case to be MMD rather than MMS. In both scenarios PTA plays a protective role in case of large vessel occlusion, and in this patient either MMD or MMS, prompt revascularization surgery was the treatment of choice.

4. Conclusions

Our case brings attention to the infrequent coexistence of ipsilateral MMD, PTA and hypoplastic vertebral and Basilar arteries, suggesting a common congenital pathogenesis, and a possible protective role of PTA in MMD. This case contributes to the scarce literature on the intriguing relationship between MMD and PTA. More studies will need to be conducted to prove that the PPTA is associated with MMD rather than only an incidental finding.

CRediT authorship contribution statement

Marilhia Cornejo: Investigation, Writing – original draft. **Ramit Singla:** Writing – review & editing. **Savdeep Singh:** Supervision. **Cheran Elangovan:** Supervision. **Balaji Krishnaiah:** Supervision.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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